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Complete primary repair of the exstrophy/ epispadias complex

Richard W. Grady, MD[a,b,*], Michael E. Mitchell, MD[a,b]

[a]Department of Urology, The University of Washington Medical Center
[b]Children's Hospital and Regional Medical Center, 4800 Sandpoint Way NE CH-81, Seattle, WA 98105, USA

Preoperative care

After delivery, the umbilical cord should be ligated with silk suture rather than a plastic or metal clamp to prevent trauma to the exposed bladder plate. A hydrated gel dressing such as Vigilon [Bard, NJ] may be used to protect the exposed bladder from superficial trauma. This type of dressing is easy to use, keeps the bladder plate from desiccating, and stays in place to allow handling of the infant with minimal risk of trauma to the bladder. Plastic wrap is an acceptable alternative. Dressings should be replaced daily and the bladder should be irrigated with normal saline with each diaper change. Other authors have advocated the use of a humidified air incubator to minimize bladder trauma [1].

The authors routinely use intravenous antibiotic therapy in the preoperative and postoperative period. This therapy decreases the chance for infection following reconstruction. The authors also perform preoperative ultrasonography to assess the kidneys and to establish a baseline examination for later ultrasonographic studies. Preoperative spinal sonographic examination should be considered if sacral dimpling or other signs of spina bifida occulta are noted on physical examination.

Operative considerations

In the newborn period, the authors perform primary exstrophy closure using general inhalation anesthesia. Nitrous oxide during primary closure may cause bowel distension, which decreases surgical exposure during the operation and increases the risk of wound dehiscence. The authors ask their anesthesiologists to avoid using it whenever possible. Some surgeons advocate the use of nasogastric tube drainage to decrease abdominal distension in the postoperative period [2]. The authors do not use nasogastric suction in most patients but do routinely use a one-time caudal block to reduce the inhaled anaesthetic requirement during the operation.

For patients older than 3 days or for newborns with a wide pubic diastasis, the authors perform anterior iliac osteotomies. Osteotomies assist closure and enhance anterior pelvic floor support, which may improve later urinary continence [3,4].

Factors that are important in the operative period include (1) use of osteotomies in selected cases and for newborn closures taking place more than 24 to 48 hours after birth, (2) ureteral stenting catheters placed intraoperatively for use in the postoperative period to divert urine, (3) avoidance of abdominal distension, and (4) use of intraoperative antibiotics [1,2].

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[*] Corresponding author.

E-mail address: richard.grady@seattlechildrens.org (R.W. Grady).

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Complete primary extrophy repair surgical technique—bladder exstrophy

After standard preparation of the surgical field, the authors place traction sutures into each of the hemiglans of the penis. These sutures should be oriented transversely. The lines of dissection are then marked (Figs. 1 and 2). Following this, 3.5 French umbilical artery catheters are placed into both ureters and are sutured in place with 5-0 chromic suture (Fig. 2). The initial dissection begins superiorly and proceeds inferiorly to separate the bladder plate from the adjacent skin and fascia. The authors use tungsten fine-tip electrocautery during this dissection. The umbilical vessels may be ligated if necessary. The periumbilical skin is incised circumferentially at this time. The umbilicus will be moved superiorly to a more anatomically normal location. It will be later used as the location to bring out the suprapubic catheter.

Penile/urethral dissection

Traction sutures are placed into each hemiglans of the penis and aid in the dissection at this point in the operation (Fig. 2). The sutures will rotate to a parallel vertical orientation (Fig. 5) as the corporal bodies rotate medially following their dissection from the "urethral wedge" (urethral plate plus underlying corpora spongiosa).

The authors begin the penile dissection along the ventral aspect of the penis as a circumcising incision (see line of dissection on Fig. 1). This step precedes the dissection of the urethral wedge from the corporal bodies because it is easier to identify the plane of dissection above Buck's fascia ventrally (Fig. 3). The Buck's fascia is deficient or absent around the corpora spongiosum. Thus, as the dissection progresses medially to separate the urethra from the corpora cavernosa, the plane shifts subtly from superficial to Buck's fascia to the plane immediately on the tunica albuginea. This change in plane must be recognized. Failure to adjust the plane of dissection will potentially extend into the corpora spongiosa, resulting in excessive bleeding during the deep ventral dissection of the urethral wedge from the corporal bodies.

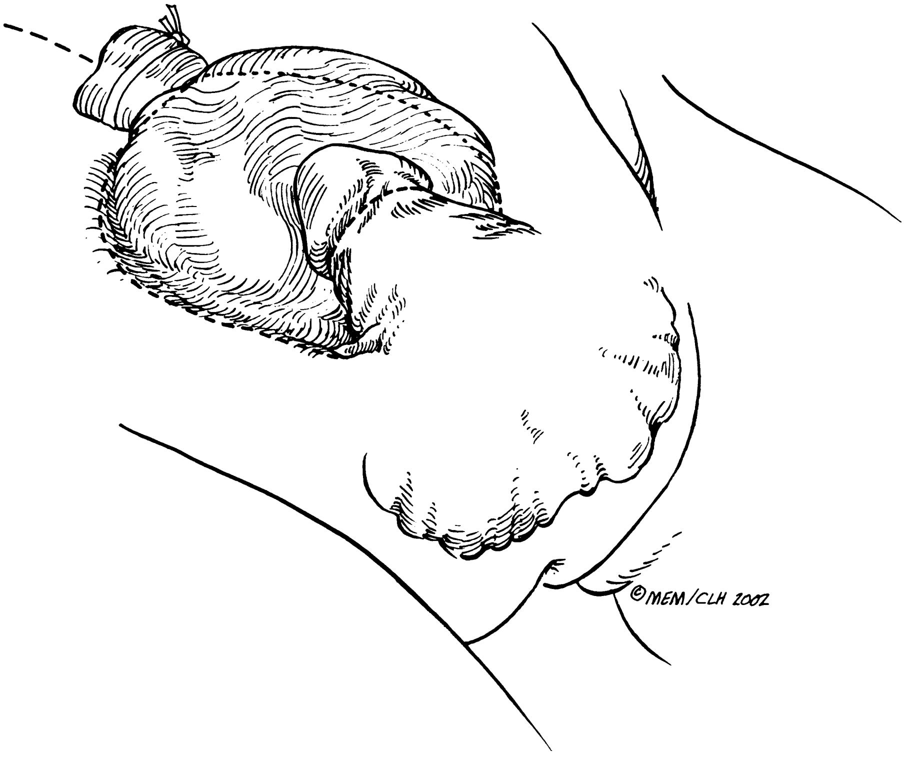

Fig. 1. Initial dissection—inferior view. Dashed lines indicate lines of dissection. Note that the lines of dissection proceed around the umbilicus and superior to it. The line of dissection also extends subcoronally around the ventral aspect of the penis. (Fig. 2 illustrates the initial lines of dissection.)

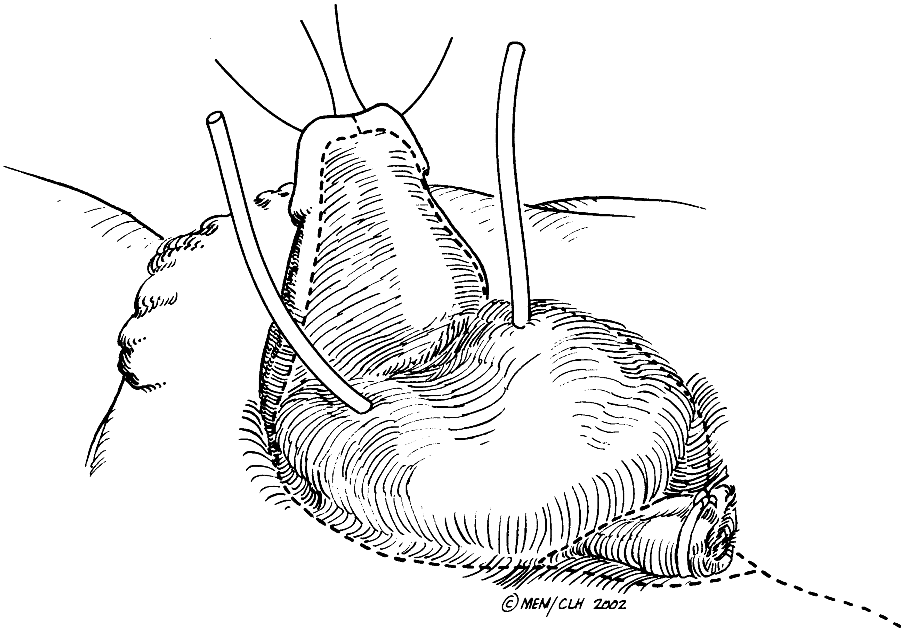

Fig. 2. View of lines of dissection from above. The urethral dissection is carried along the lateral aspect of the urethral plate.

The authors apply methylene blue or brilliant green to the urethra to help identify the demarcation of urothelium and squamous epithelium. Injection of the surrounding tissues with 0.25% lidocaine and 1:200,000 U/mL epinephrine also improves hemostasis, assisting the dissection. The margins of the dorsal urethra are usually obvious. Shallow incisions are made laterally to begin the urethral dissection (Fig. 4). Sharp dissection is required to develop the plane between the urethral wedge and the corpora cavernosa. Careful dissection will preserve urethral width.

Careful lateral dissection of the penile shaft skin and dartos fascia from the corporal bodies will avoid damage to the laterally located neurovascular bundles contained in Buck's fascia and oriented on the lateral aspect of corpora cavernosa of the epispadic penis. The lateral dissection on the penis thus must be superficial to Buck's fascia because of the lateral location of the neurovascular bundles in the epispadic penis.

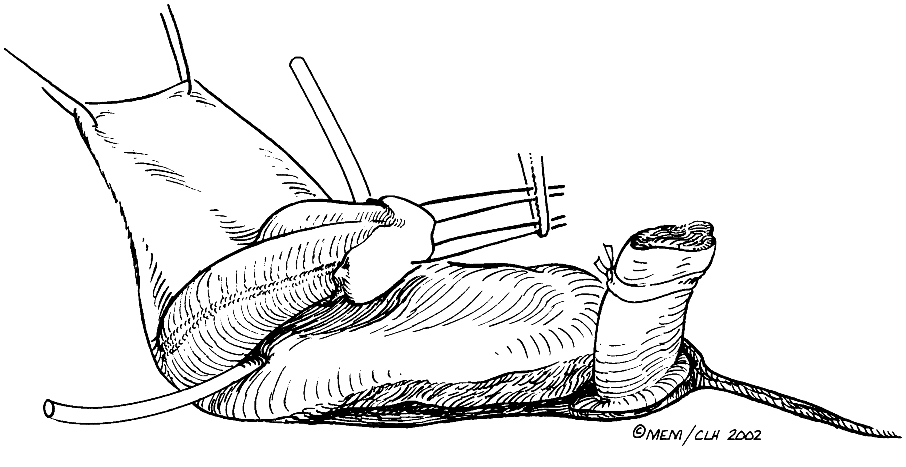

Fig. 3. Ventral dissection is initiated most easily below the glans with a circumcising incision. The dissection may be carried proximally.

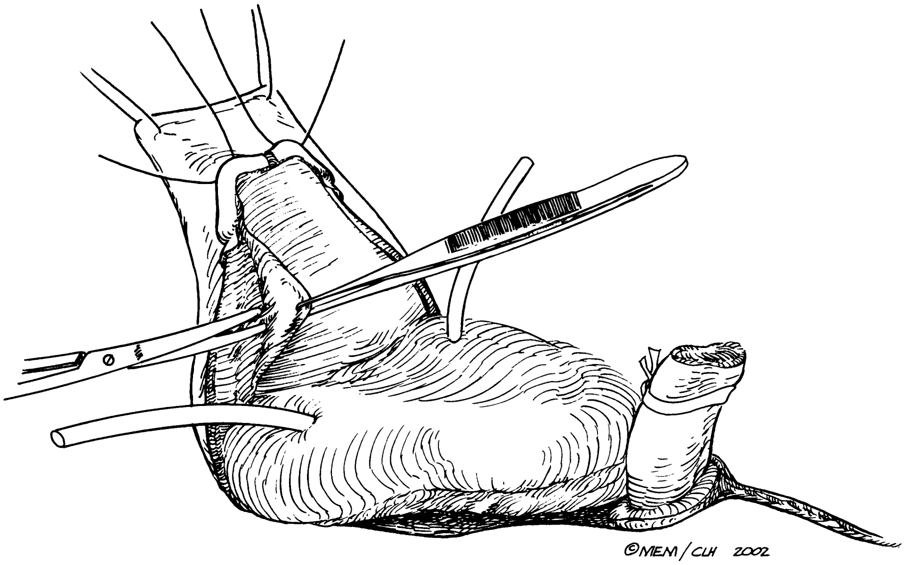

Fig. 4. The urethra is dissected from the corporal bodies. This plane of dissection is developed from both a ventral and lateral perspective using sharp dissection.

Complete penile disassembly and deep dissection

After a plane is established between the penis and the urethral wedge (Fig. 5), the penis may be disassembled into three components: the right and left corporal bodies with their respective hemiglans, and the urethral wedge (urothelium with underlying corpora spongiosa) [5]. Disassembly facilitates exposure (later in the operation) of the intersymphyseal band and allows adequate proximal dissection. The authors have found the easiest plane of dissection between

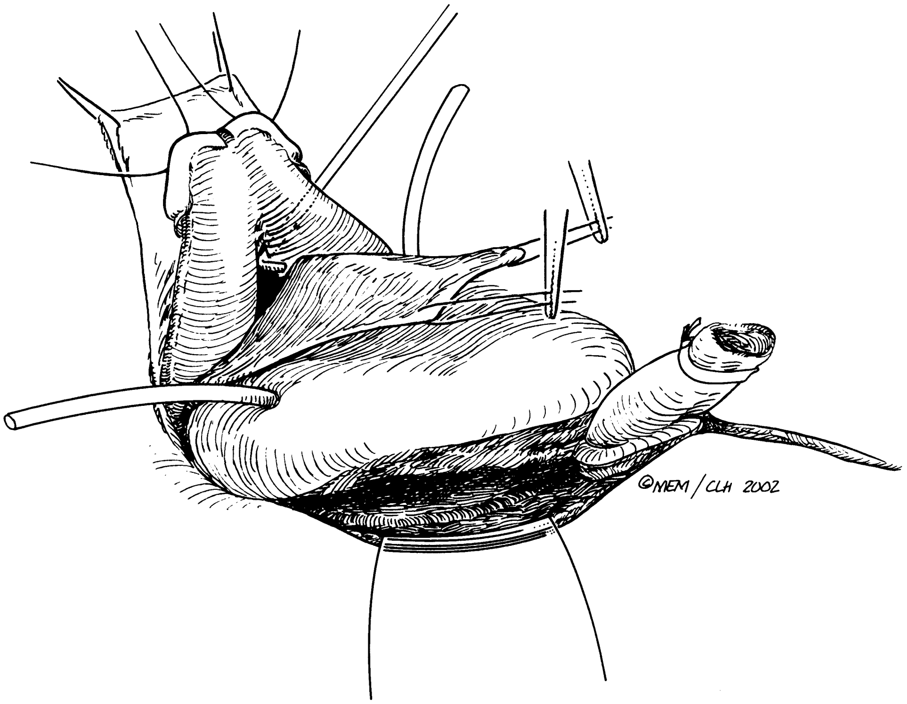

Fig. 5. Disassembly of the corpora cavernosa from the urethra and corpora spongiosum (the urethral wedge) can often be most easily begun at the position depicted here. The dissection is carried distally to completely separate the glans penis.

the corporal bodies to be proximal and ventral (Fig. 6). The plane of dissection here is immediately on the level of the tunica albuginea on the corpora cavernosa. After the plane is established between the urethral wedge and the corpora cavernosa, this dissection is carried distally to separate the three components from each other (Fig. 7). Complete separation of the corporal bodies facilitates exposure to the pelvic diaphragm for deep dissection. The corporal bodies may be completely separated from each other because they have unique and separate blood supplies (Figs. 6 and 7). It is important to dissect the underlying corpora spongiosum with the urethra; the blood supply to the urethra is based on this tissue, which will appear wedge shaped after the dissection from the adjacent corpora cavernosa. The urethral/corporus spongiosal component will later be tubularized and transposed ventral to the corpora cavernosa. Paraexstrophy skin flaps cannot be used with this technique because their dissection will devascularize the distal urethra. Because the bladder and urethra are moved posteriorly in the pelvis as a unit (with a common proximal blood supply), division of the urethral wedge is not an option. In some cases, a male patient will be left with a "hypospadias" requiring later surgical reconstruction.

After separating the components distally (Fig. 7), the urethral dissection is extended proximally to the level of the bladder neck. Complete separation of the urethra and corporal bodies creates sufficient exposure to allow the deep dissection of the intersymphyseal band required to move the bladder and urethra posteriorly (Figs. 6 and 7). Medially, under the

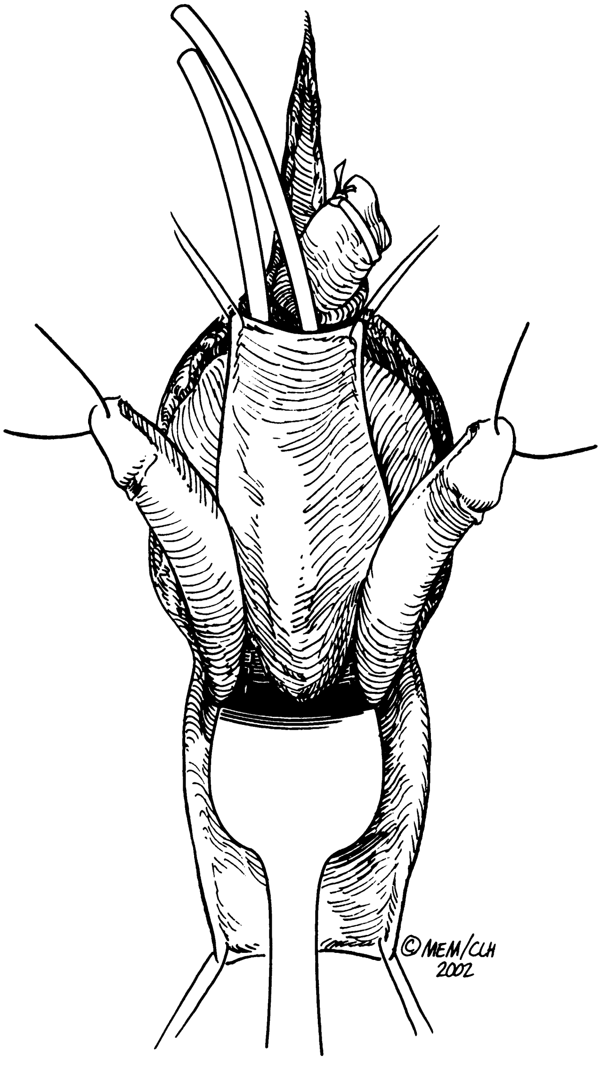

Fig. 6. Perineal view of complete disassembly. Notice depth of dissection.

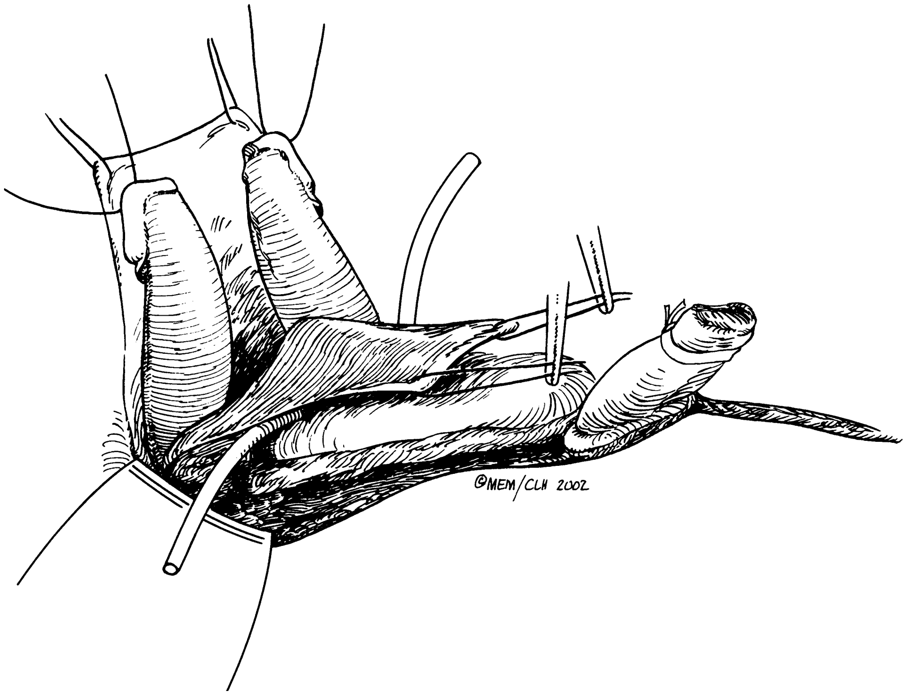

Fig. 7. The urethra and corpora cavernosa are separated distally to allow deep dissection of the pelvic floor musculature.

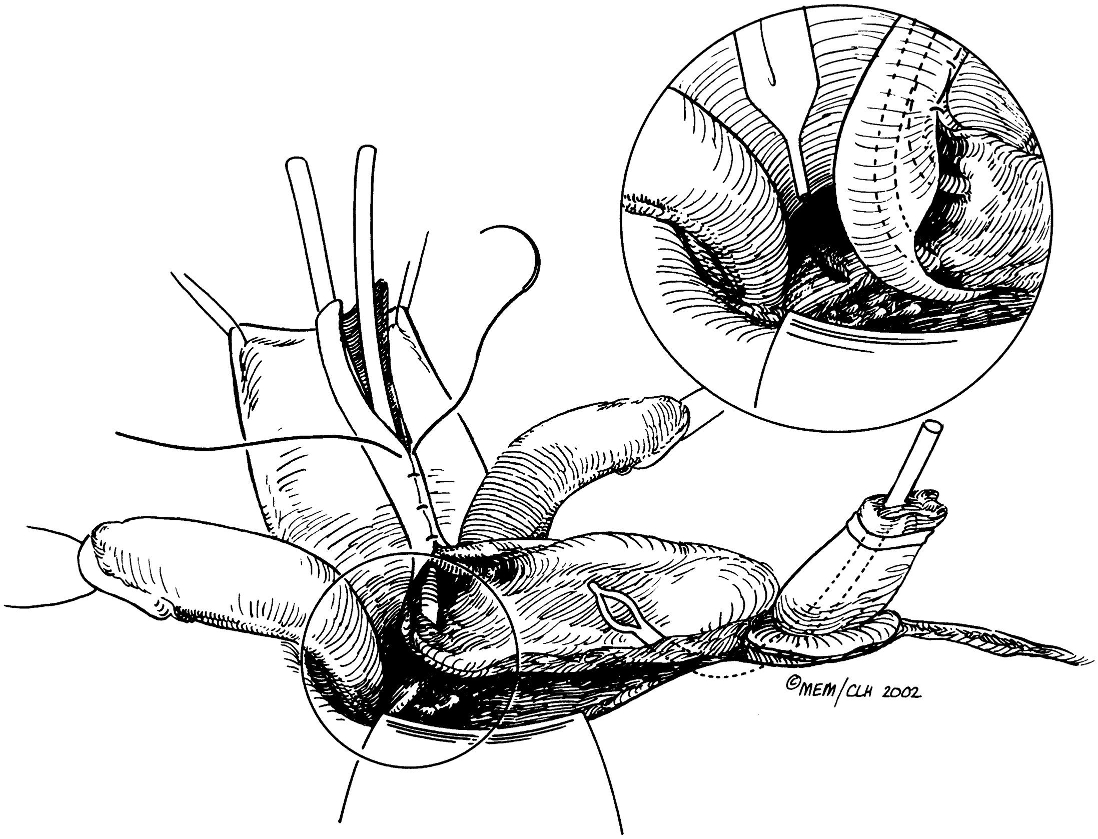

Fig. 8. The exposure for deep dissection is optimal after complete separation of the corpora cavernosa. It is crucial to adequately divide the intersymphyseal band (shown in inset) to allow the bladder and urethra to move posteriorly.

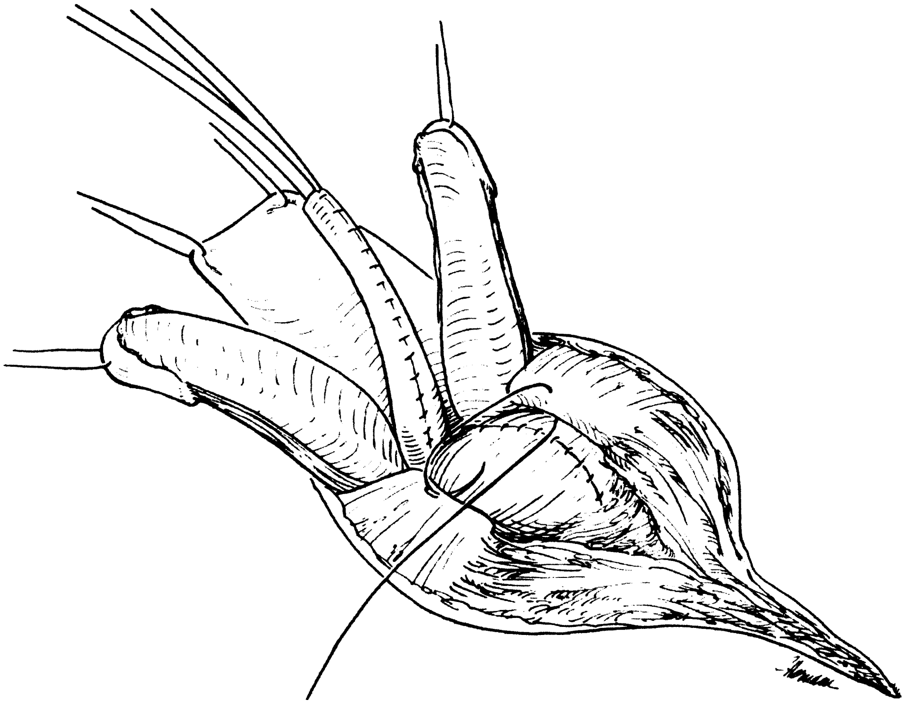

Fig. 9. The urethra and bladder are reapproximated in a two-layer closure.

urethral wedge, the plane of dissection is immediately on the tunica albuginea of the corpora cavernosa (Fig. 6).

Proximal dissection of the urethral wedge from the corporal bodies provides the exposure critical to the posterior placement of the bladder neck and proximal urethra. Dissection along the medial aspect of the corpora cavernosa leads to the intersymphyseal band (the anterior aspect of the pelvic diaphragm). Deep incision of the intersymphyseal band, posterior and lateral to each side of the urethral wedge, is absolutely necessary to allow the bladder to achieve a posterior position in the pelvis (Fig. 8, see inset) This dissection should be extended until the pelvic floor musculature becomes visible. Computerized tomography of the pelvis demonstrates that the intersymphyseal band lies posterior to the bladder neck and urethra. Failure to adequately dissect the bladder and urethral wedge from these surrounding structures will prevent posterior movement of the bladder in the pelvis and thus create anterior tension along the urethral plate.

Primary closure

After the deep dissection is performed and the bladder and urethral wedge are adequately freed from the surrounding tissues, the majority of the procedure is done and the closure is straightforward and anatomic. Prior to reapproximating the bladder, a suprapubic tube is placed and brought out through the umbilicus. A primary closure of the bladder is then performed using a three-layer closure with monofilament absorbable suture (ie, Monocryl and Vicryl; Ethicon, Cornelia, GA). The urethra is tubularized using a two-layer running closure with monofilament and braided absorbable suture (Fig. 9). The tubularized urethra is then transposed ventral to the corpora cavernosa in an anatomically normal position.

The pubic symphysis is reapproximated using 2-0 polydiaxonone interrupted sutures. Knots are placed anteriorly to prevent suture erosion into the bladder neck (Fig. 10). The rectus fascia is reapproximated using an interrupted or running 2-0 polydiaxonone suture. Interrupted sutures are also placed to reapproximate the corpora cavernosa (Fig. 11). The penile shaft skin is reconfigured using either a primary dorsal closure or reversed Byars flaps, if needed, to provide dorsal skin coverage. The abdominal wall skin is reapproximated using a two-layer closure of absorbable monofilament suture.

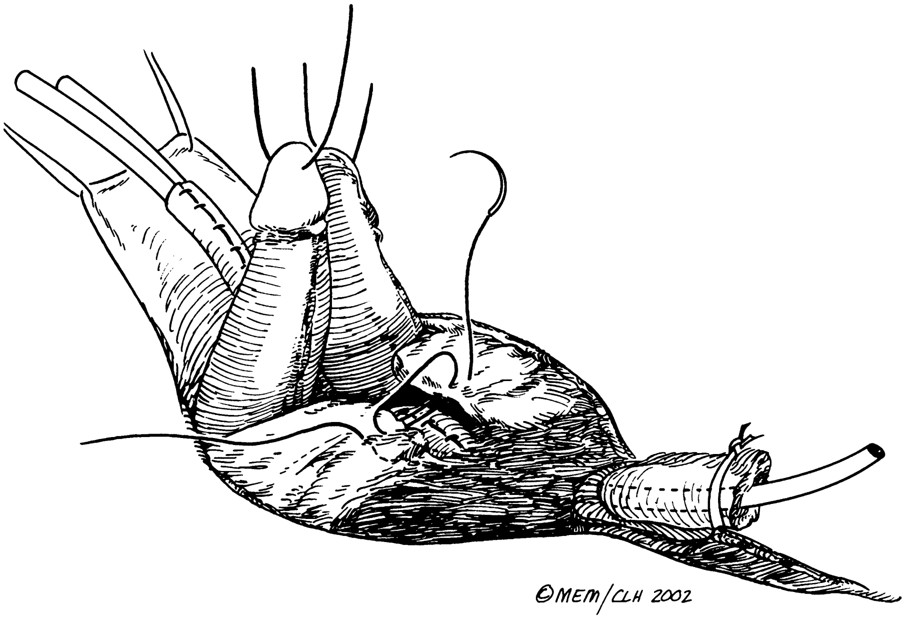

Fig. 10. After the bladder and urethra are reconstructed, these structures will move posteriorly. The urethra will assume a more normal anatomic position. The pubis symphysis is reapproximated with two figure-eight sutures.

The corpora cavernosa will tend to rotate medially with closure as the lateral margins of Buck's fascia of the corpora cavernosa are approximated (Fig. 10). This maneuver assists in correcting the dorsal deflection and is readily observed because the vertical lie of the previously horizontally placed glans traction sutures is appreciated. Occasionally, significant discrepancies in the dorsal and ventral aspects of the corpora cavernosa will require placement of a dermal graft. This is rarely needed, however, in the newborn closure.

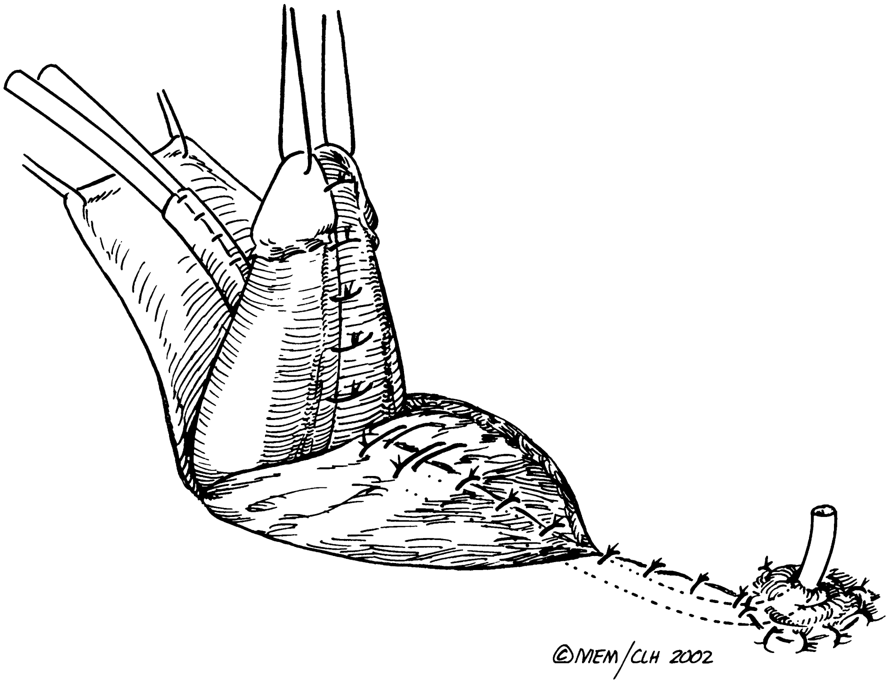

Fig. 11. The corporal bodies will rotate medially so that the neurovascular bodies are located medially. The suprapubic tube can be brought out through the umbilicus. The umbilicus is moved superiorly to a more normal anatomic location.

The urethra can then be approximated to each hemiglans ventrally, creating an orthotopic meatus (Fig. 11). The glans is reconfigured using interrupted mattress sutures of polydiaxonone suture followed by horizontal mattress sutures of 7-0 monofilament suture to reapproximate the glans epithelium. The neomeatus is matured with 7-0 braided polyglactin suture as with the authors' standard hypospadias repair. When needed, the authors will also debulk the ventral glans to create a conical-appearing glans. Tacking sutures are place ventrally and dorsally to prevent the penile shaft skin from riding over the corporal bodies, "burying" the penis (Figs. 12 and 13).

In roughly half of the authors' cases, the urethra lacks enough length to reach the glans. In this situation, the urethra may be matured along the ventral aspect of the penis to create a hypospadias, which is corrected at a later date as a second-stage procedure. The authors often leave the ventral shaft skin redundant in these patients to assist in later penile reconstructive procedures.

Postoperative care

The patient is immobilized to decrease lateral stresses on the closure. A number of options exist for this purpose. The authors prefer to use a spica cast for immobilization. A spica cast for 3 weeks prevents external hip rotation and optimizes pubic apposition, thus facilitating early discharge and home care (Fig. 14). Modified Buck's traction has been used by many groups for a period of 3 to 4 weeks. A posterior lightweight splint can be used in newborns when the child is out of traction to facilitate home care and early removal of traction. The authors have stopped using Buck's traction because spica casts are easier for the families to care for at home. External fixation devices have also been advocated by several centers. Fixator pins for these devices should be cleaned several times a day to reduce the chance of infection. Internal fixation may be necessary in older patients.

Because of the high incidence of vesico-ureteral reflux, the authors prescribe low-dose suppressive antibiotic therapy for all newborns after bladder closure. This is continued until the vesico-ureteral reflux is corrected or it resolves spontaneously. Postoperative factors that appear

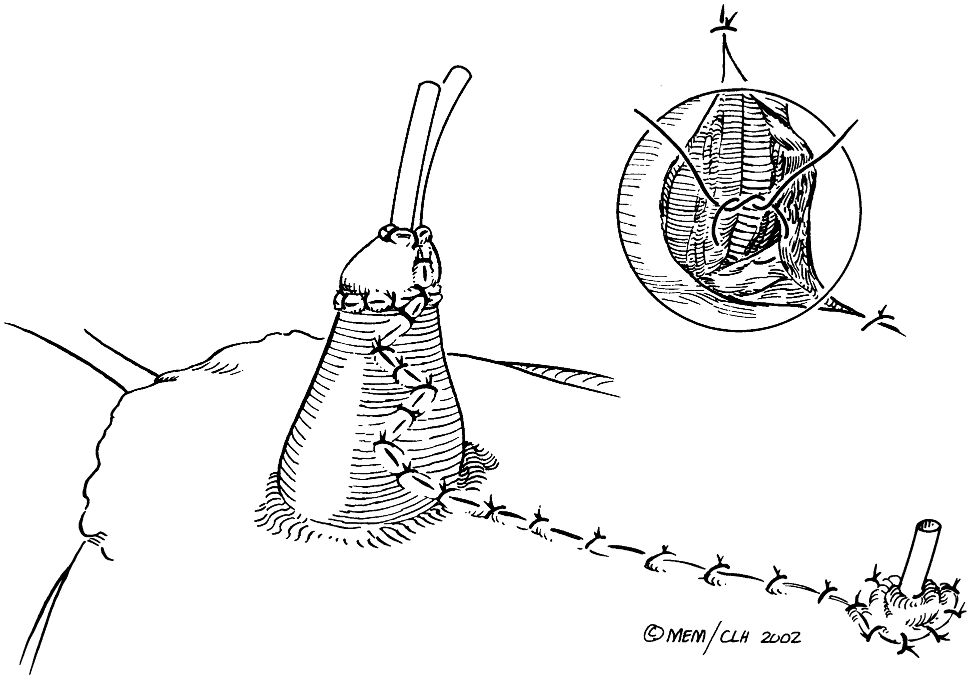

Fig. 12. To adequately cover the penis dorsally, z-plasty incisions may be necessary. Tacking sutures are also employed dorsally (see insert).

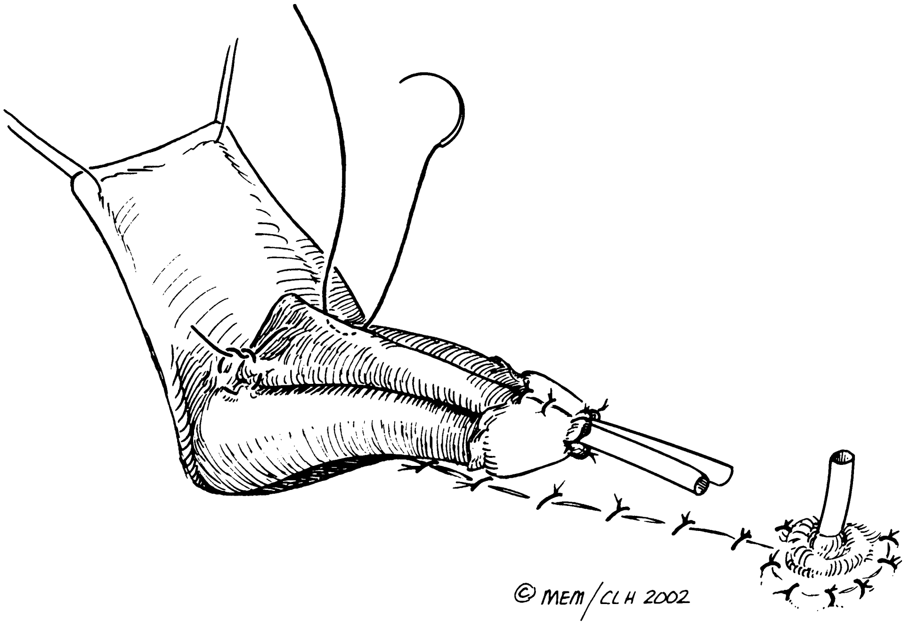

Fig. 13. The ventral shaft skin is secured to the base of the penis to prevent the penile shaft skin from riding over the body of the penis. The ureteral catheters are brought out through he urethra.

to directly impact the success of initial closure include (1) postoperative immobilization, (2) use of postoperative antibiotics, (3) ureteral stenting catheters, (4) adequate postoperative pain management, (5) avoidance of abdominal distension, (6) adequate nutritional support, and (7) secure fixation of urinary drainage catheters [1].

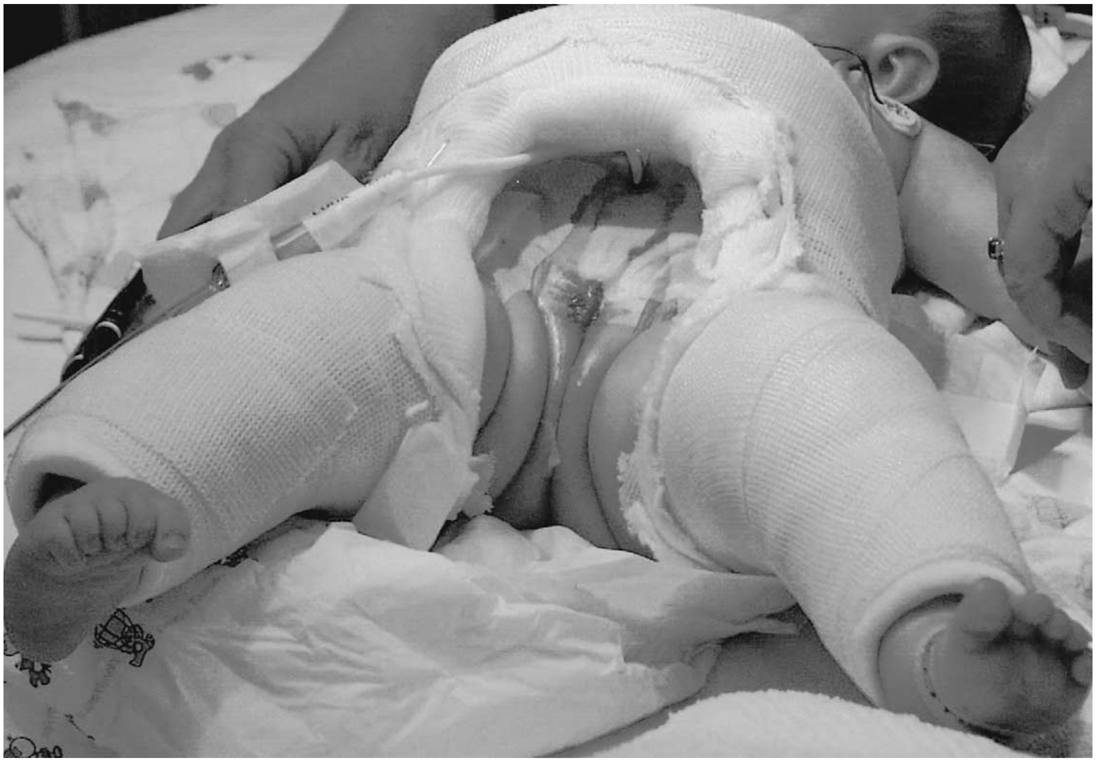

Fig. 14. Use of a spica cast postoperatively to immobilize the pelvis and prevent hip abduction.

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